Role of Dental Findings in the Diagnosis of Idiopathic Hypoparathyroidism

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CASE REPORT

ABSTRACT

Idiopathic hypoparathyroidism (IHP) is a rare endocrinopathy, characterized by the disturbances in the calcium and phosphorous metabolism, owing to deficiency in parathyroid hormone, which leads to tetanic manifestations. Onset of the clinical features occurs early in the life and the severity depends on the extent of chemical imbalance.

This article describes a case of 22-year-old male patient undiagnosed for 12 years with this endocrinopathy (IHP). Overretained deciduous teeth, delayed eruption, impacted tooth and short roots probably resulting from untreated hypocalcemia during the developmental phase of dentition enabled us to unearth this endocrinopathy through a series of investigations. Thus, the article emphasizes the importance of dental findings of this endocrinopathy.

Keywords: Idiopathic hypoparathyroidism, Hypocalcemia, Hyperphosphatemia, Tetany, Hypoplasia, Stunted roots.

INTRODUCTION

Parathyroid hormone released from parathyroid glands plays an essential role in homeostasis of calcium and phosphate and in turn influencing upon the normal mineralization of bone and teeth as well as in maintenance of neuromuscular activity.1 Hyposcretion of this hormone leads to a relatively rare metabolic disorder known as hypoparathyroidism, which is characterized by hypocalcemia and hyperphosphatemia.2,3 Hypoparathyroidism may occur as follows:

a. In newborns as a result of mother’s hypoparathyroidism states.
b. As a result of excision of parathyroid gland during thyroidectomy or
c. May develop as an isolated entity of unknown etiology, called idiopathic hypoparathyroidism (IHP).1,4

Symptoms of IHP usually appear during the first decade of life but may become evident at any age.1 In acute form, it causes hypocalcemia with consequent paresthesia, muscular spasm and seizures. Long standing cases manifest with visual impairment from cataracts.3 Dental manifestations comprises of enamel hypoplasia, widened pulp chambers, pulp stones, shortened roots, delayed eruption and hypodontia.5,6

To the best of our knowledge since 1966, only six studies have been published reporting the dental anomalies in IHP patients.1,3,4,7-9 This article describes the importance of analyzing the clinical presentation with dental manifestations, which lead us to identify an endocrinopathy in a 22-year-old young adult, undiagnosed for about 12 years.

CASE REPORT

A 22-year-old male reported to the department of oral medicine and radiology, seeking an advice regarding absence of some teeth. Family history was unremarkable. He had, however discontinued his education as he could not cope up with his peer group. On further inquiry patient’s father reported subject’s delayed milestones. His medical history revealed that he had suffered from epileptic seizures at the age of 11 years for which he was treated with anticonvulsant drugs clonazepam and carbamazepine. Seizures persisted for about 2 to 3 years in spite of medicines. A year ago, he was diagnosed with bilateral cataracts which were surgically treated.

Patient was of short stature and was subnormal in his alertness and response to questions. An intraoral examination revealed normal soft tissues. However, the dentition revealed diastema in the upper and lower arch, multiple retained deciduous teeth (#53, #63, #64 and #65) and missing permanent teeth (#13, #14, #17, #18, #23, #24, #25, #27, #34, #37, #38, #42, #47 and #48) (Fig. 1). Panoramic and periapical radiographs showed multiple impacted teeth, shortened roots...
in the premolars, retarded root development in the second molars and microdontia associated with enamel hypoplasia in the impacted third molars (Figs 2 and 3). There were no gross changes in the bone trabecular pattern. Based on the history, clinical and radiological findings, the possibility of endocrine disturbance pertaining to thyroid, pituitary, parathyroid and vitamin D deficiency state was thought of as differential diagnosis.

The patient was subjected for hematological and serological analysis. Hematological studies showed normal readings. Thyroid panel by chemiluminescence disclosed T3 of 155.6 ng/dl (normal, 60-180 ng/dl), T4 of 8.4 μg/dl (normal, 4.5-12.60 μg/dl), TSH of 1.52 μIU/ml (normal, 0.35-5.5 μIU/ml) and growth hormone was slightly on a higher level of 1.1 ng/ml (normal, 0.01-1 ng/ml). However, chemical analyses revealed serum calcium of 5.1 mg/dl (normal, 9-11 mg/dl) and serum phosphate of 6.3 mg/dl (normal, 3-5 mg/dl) with alkaline phosphatase being in normal range (118 IU).

History of tetanic spasms (possibly misdiagnosed earlier as seizures), cataracts, radiographic details and the picture of hypocalcemia and hyperphosphatemia with normal thyroid and growth hormone assays and normal bone morphology prompted us to think about the possibility of parathyroid gland disturbance.

An endocrinologist opinion was taken to rule out hypoparathyroidism or pseudohypoparathyroidism as the possible cause for the patient’s systemic and dental manifestations. Examination of the patient’s hands and limbs was carried out to rule out pseudohypoparathyroidism, which reveals short metacarpals and metatarsals. Hand-wrist radiographs revealed normal findings. Further, serum parathyroid hormone assay was carried out with a reported value of <2.50 pg/ml (normal, 14-72 pg/ml). Finally, based on chemical analysis and radiographic features, a diagnosis of hypoparathyroidism was made and was confirmed with Ellsworth-Howard test in which 250 units of parathion was injected IM and urinary phosphorous level was increased as much as ten times in 5 hours. Patient was advised calcium gluconate (24 μg/day) and calcitrol (1 μg/day). After 1 month, calcium levels were slightly increased along with decreased levels of phosphate.

DISCUSSION

Clinical presentation of hypocalcemia, hyperphosphatemia along with dental changes is the sequelae of parathyroid hormone deficiency (hypoparathyroidism) or inability of the target organs (kidneys and bones) to respond to this hormone leading to pseudohypoparathyroidism (PTH). PTH is a rare metabolic disorder with clinical findings as similar as IHP. Distinction is made by the serum parathyroid hormone levels and by testing renal resistance to exogenous hormone (Ellsworth-Howard test).

The most common presenting symptom of IHP is tetany, which occurs as a result of low serum level of calcium which if left untreated causes increased neuromuscular activity. This could be the possible cause in the present case resulting in tetany which was mistaken and treated as epilepsy for about 12 years. Hence, even with the treatment of antiepileptic drugs, patient suffered with tetanic spasms.
Ectodermal disorders are the most common in the patients with IHP. These disorders include scaling of the skin, deformities of the nails, opacities of the cornea and lens, keratoconjunctivitis and dental abnormalities.3

Dental findings in the previous six studies comprised of enamel hypoplasia as the most frequent finding, however, short rounded roots, hypodontia, lack or delayed tooth eruption, partial anodontia and microdontia were also noted,5,6,10 of which most of the findings were observed in the present case.

Evidence from the previous studies emphasized that dental abnormality in IHP was the result of calciotraumatic response which coincides with the age of onset and reflect periods of hypocalcemia during the tooth development11,12. In the present case, the chemical imbalance which might have occurred at around the age of 10 to 11 years presented with enamel hypoplasia and microdontia with respect to third molars (mean age of calcification for third molars), impairment of root development of the premolars, second molars and third molars. These manifestations have been attributed to the disturbance in mineralization, alterations in the formation of Hertwig’s epithelial root sheath coupled with other ectodermal disorders, lack of differentiation of odontoblasts or due to resorptive processes.1

As the patients with this endocrinopathy can land up in dental clinic with a complaint of discolored (hypoplastic) teeth or delayed eruption of the teeth, treatment and the prognosis will remain uncertain unless the root cause is diagnosed.

So, thorough evaluation of the dental and clinical findings aided with relevant investigations can help in the early detection of this endocrine-deficiency syndrome. Hence, prompt intervention during the early stages of the disease cannot only minimize the dental anomalies but also improve the overall health of the patient.

REFERENCES